COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY (CSID)

Symptoms, Diagnosis, and Treatment

FOR PEDIATRIC GASTROENTEROLOGISTS

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Financial Disclosures

- [Disclose financial relationships with manufacturers and medical organizations here (e.g., QOL Medical, LLC); if none, list "None."]

WHAT IS CSID?

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

CSID: Congenital Sucrase-Isomaltase Deficiency

Sucrase-Isomaltase

- An enzyme that digests the majority of dietary carbohydrates
- Table sugar (sucrose) and many starches (e.g., potatoes, bread)
- Expressed in the microvilli of the brush border membrane
- Releases glucose and fructose from sucrose (sugar) so they can be absorbed into the bloodstream

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Congenital Sucrase-Isomaltase Deficiency

The first report of an autosomal recessive Congenital Sucrase-Isomaltase Deficiency (CSID) was published in 1960.

Diarrhoea Caused by Deficiency of Sugar-Splitting Enzymes


THE LANCET

VOLUME TWO

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Weijers HA, Van De Kamer JH, Mossel DAA, Dicke WK. Diarrhoea Caused by Deficiency of Sugar-Splitting Enzymes. *Lancet*. 1960;276(7145):296-7.

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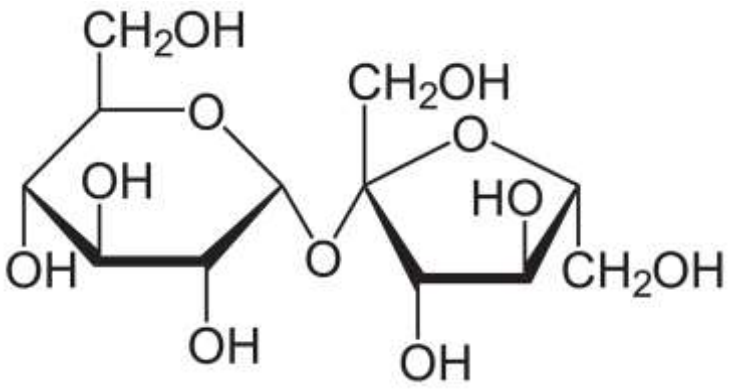
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

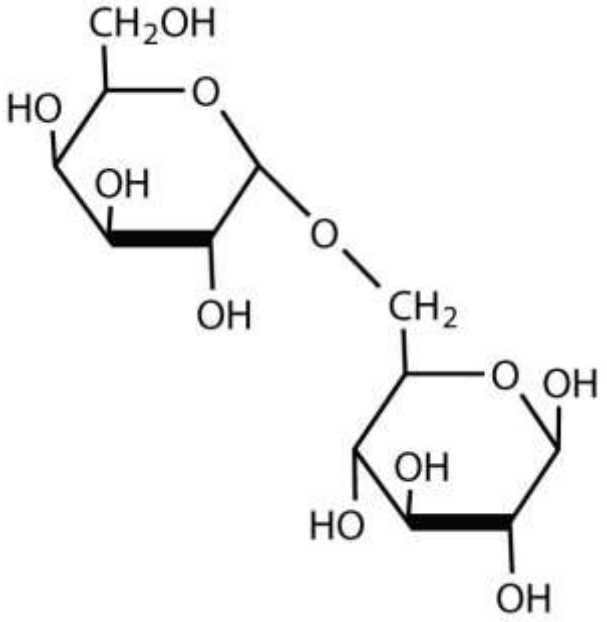
Sucrase-Isomaltase Substrates

Sucrose



Glucose + Fructose
(α–1,2 glycosidic bond)

Isomaltose



Glucose + Glucose
(α–1,6 glycosidic bond)

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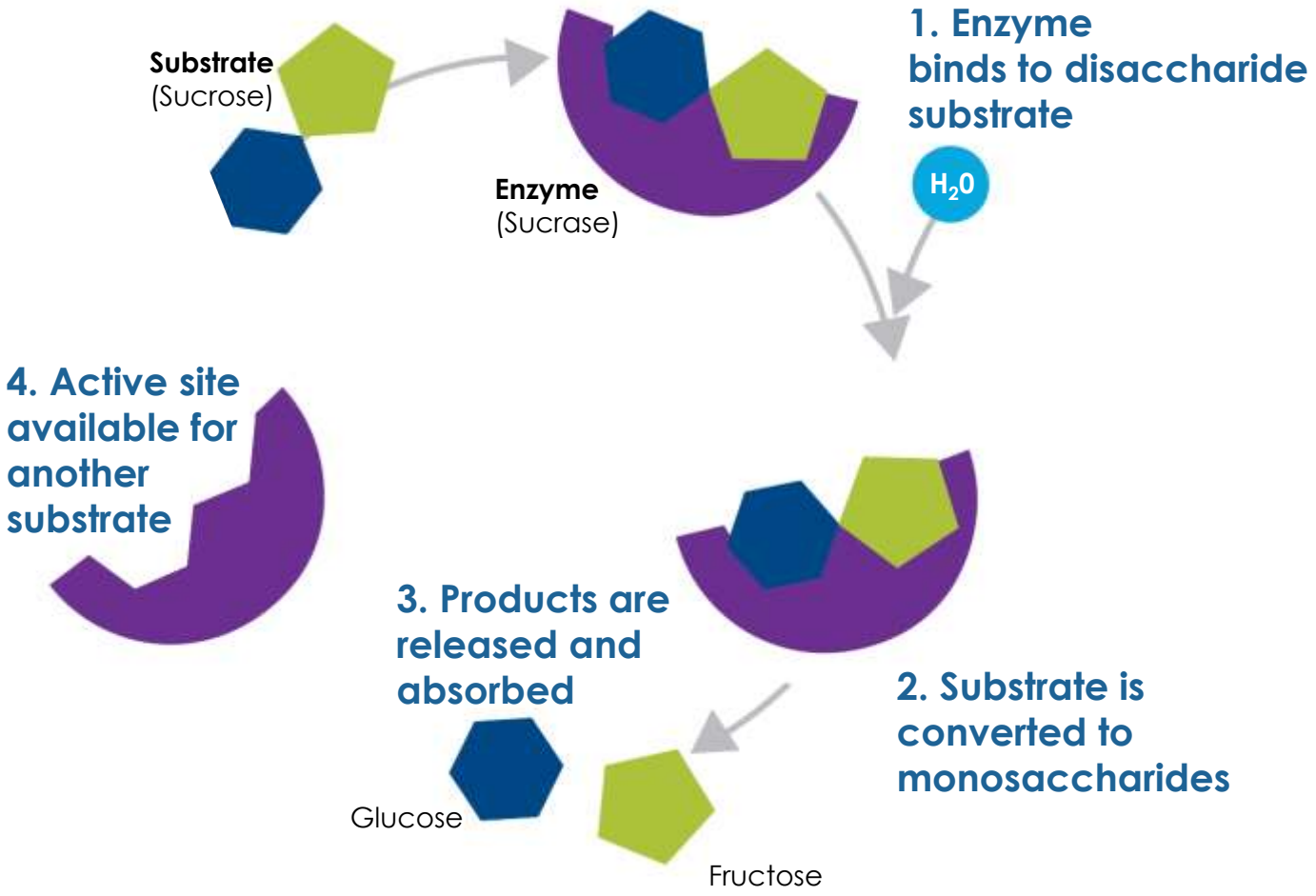
How Sucrase Works to Hydrolyze Sucrose



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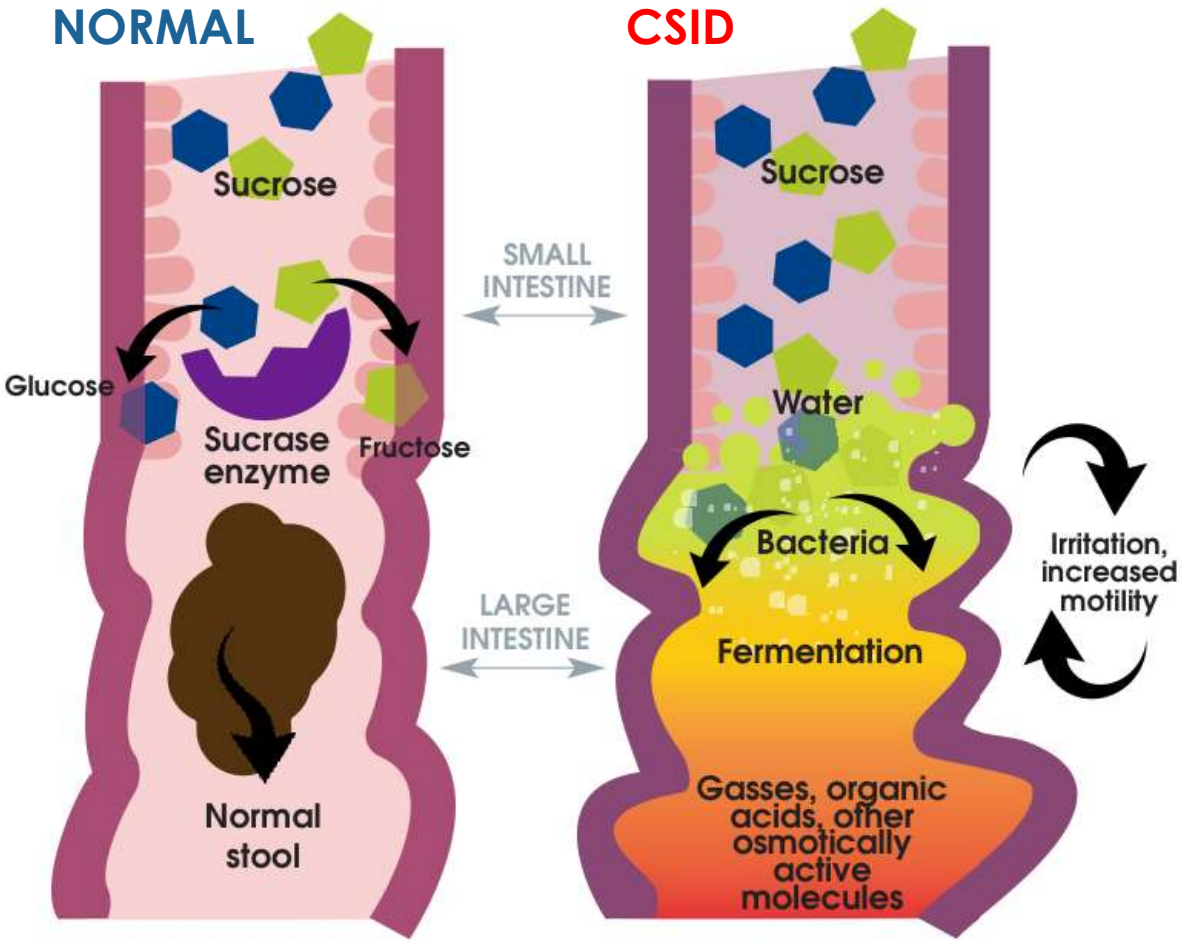
CSID Carb Maldigestion Pathophysiology



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SI Gene

- Encodes a heterodimer with 2 active sites – sucrase and isomaltase
  - Located on chromosome 3[1]
  - Very large – approximately 100 kilobases[1]
  - 48 exons encoding 1827 amino acids[1]
- 2146 rare variants with 880 *SI* rare pathogenic variants (*SI*-RPVs)[2]
  - Sucrase-Isomaltase protein transported and anchored to apical membrane of enterocytes to digest disaccharides in intestinal lumen[3]

1. Uhrich S, Wu Z, Huang JY, Scott CR. Four Mutations in the SI Gene Are Responsible for the Majority of Clinical Symptoms of CSID. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S34-5.
2. Garcia-Etxebarria K, Zheng T, Bonfiglio F, et al. Increased Prevalence of Rare Sucrase-Isomaltase Pathogenic Variants in Irritable Bowel Syndrome Patients. *Clin Gastroenterol Hepatol.* 2018;16(10):1673-76.
3. Naim HY, Heine M, Zimmer KP. Congenital Sucrase-Isomaltase Deficiency: Heterogeneity of Inheritance, Trafficking, and Function of an Intestinal Enzyme Complex. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S13-20.

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Role of *SI* Gene

- CSID – hypomorphic variants result in lack of active protein at the cell surface causing reduced digestion, malabsorption, and colonic fermentation of carbs[1]

CSID Genotypes

- In 31 biopsy-diagnosed CSID patients, at least 1 of the 4 most common CSID variants was present in approximately 60% of the patients' alleles[2]
  - Genotypes include homozygous, compound heterozygous, and simple heterozygous

1. Naim HY, Heine M, Zimmer KP. Congenital Sucrase-Isomaltase Deficiency: Heterogeneity of Inheritance, Trafficking, and Function of an Intestinal Enzyme Complex. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S13-20.
2. Uhrich S, Wu Z, Huang JY, Scott CR. Four Mutations in the SI Gene are Responsible for the Majority of Clinical Symptoms of CSID. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S34-5.

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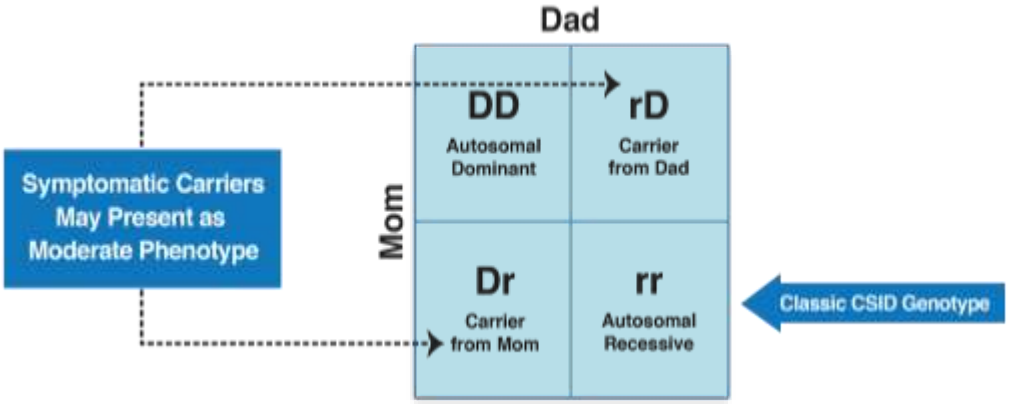
CSID Genotypes and Phenotypes



- **CSID originally assumed to be an autosomal recessive disease[1]**
  - Sucrase activity is very low (<15 µM/min/g); most severe symptoms
- **Recent studies also found symptomatic disease in heterozygotes[2]**
  - Sucrase activity low (<25 µM/min/g)
  - Correlations between CSID phenotype and heterozygous genotype

1. Chumpitazi BP, Robayo-Torres CC, Opekun AR, Nichols BL Jr, Naim HY. Congenital Sucrase-Isomaltase Deficiency: Summary of an Evaluation in One Family. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S36.
2. Uhrich S, Wu Z, Huang JY, Scott CR. Four Mutations in the SI Gene are Responsible for the Majority of Clinical Symptoms of CSID. *J Pediatr Gastroenterol Nutr.* 2012;55(suppl 2):S34-5.

HOW DO CSID PATIENTS PRESENT?

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CSID Has Three Different Presentations in Pediatrics

- Infants
- Toddlers
- Adolescents

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CSID Signs and Symptoms in Infants

- Usually a more clinically-significant phenotype
- Asymptomatic for the first 4 to 6 months
- Non-bloody, non-mucus diarrhea develops after introduction of fruits
- Bloating usually present
- Crampy abdominal pain and colic

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CSID Signs and Symptoms in Infants

- Significant diaper rash present due to acidic diarrhea
- Symptoms better when baby is fasting
- Poor weight gain or failure to thrive

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CSID Signs and Symptoms in Toddlers

- Intermittent diarrhea (malabsorption)
- Crampy abdominal pain
- Bloating and flatulence
- Aversion to fruits and sweets
- Better if fasting

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

CSID Signs and Symptoms in Adolescents

- Mild cases may present with only bloating or flatulence
- More severe cases may present with IBS-D-like symptoms
- What happens if you have constipation and CSID at the same time?
- Increased prevalence of rare sucrase-isomaltase genetic variants in irritable bowel syndrome patients suggests some of these patients may have been misdiagnosed[1]

1. Garcia-Etxebarria K, Zheng T, Bonfiglio F, et al. Increased Prevalence of Rare Sucrase-isomaltase Pathogenic Variants in Irritable Bowel Syndrome Patients. *Clin Gastroenterol Hepatol.* 2018;16(10):1673-76.

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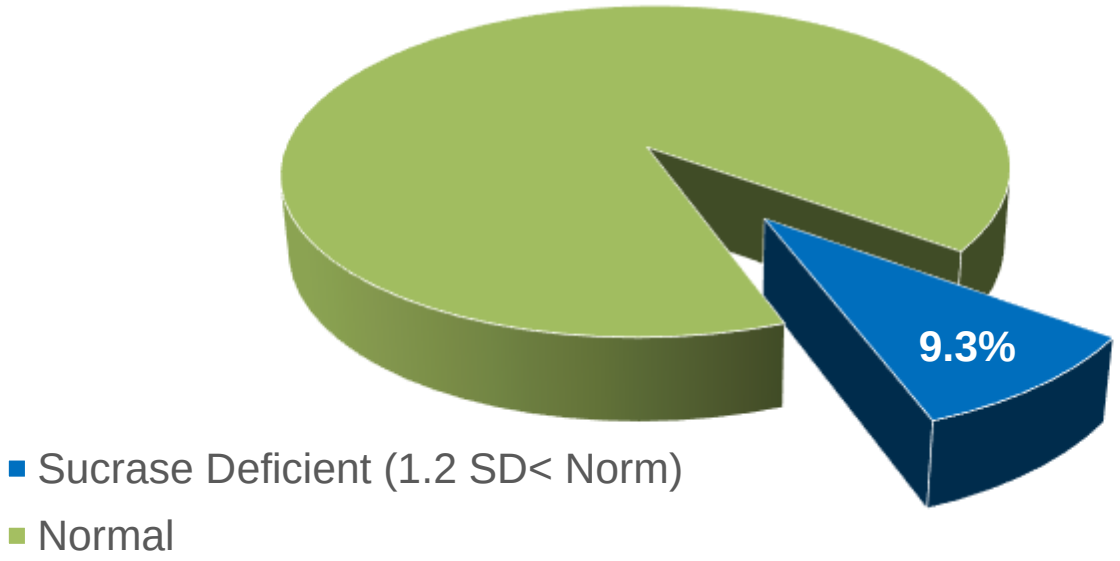
How Common Is Sucrase Deficiency?

Study of Idiopathic Sucrase Deficiency[1]
N = 27,875



1. Nichols BL Jr, Adams B, Roach CM, Ma CX, Baker SS. Frequency of Sucrase Deficiency in Mucosal Biopsies. *J Pediatr Gastroenterol Nutr*. 2012;55 (suppl 2):S28-30.

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Incidence of Sucrase-Isomaltase Rare Pathogenic Variants (*SI*-RPV) in GI Patients

	Chronic Diarrhea[1]	IBS-D[2]
Subjects (N)	**308**	**952**
Known CSID Variants (n)	**14**	**40**
Incidence	**4.5%**	**4.2%**

- *SI*-RPV does not always cause CSID, but highly correlated
- Data suggests a portion of CSID patients might be misdiagnosed with IBS-D
- IBS-D symptoms are very similar to CSID symptoms
- Consider CSID in your diagnostic algorithm *especially* if patient is unresponsive to low-FODMAP diet/IBS-D treatments

1. QOL Medical, LLC. Data on file.
2. Garcia-Etxebarria K, Zheng T, Bonfiglio F, et al. Increased Prevalence of Rare Sucrase-Isomaltase Pathogenic Variants in Irritable Bowel Syndrome Patients. *Clin Gastroenterol Hepatol.* 2018;16(10):1673-76.

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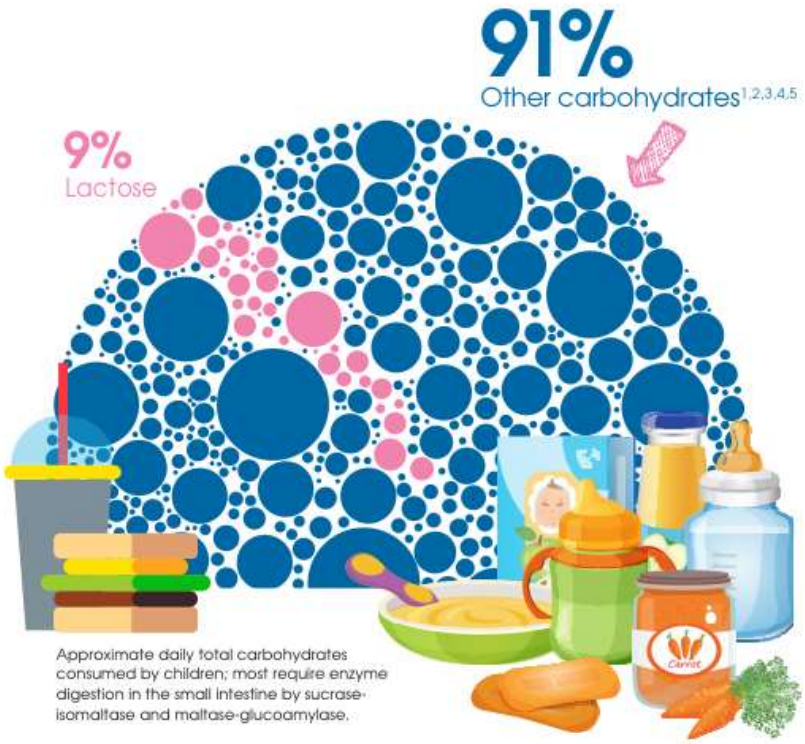
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

it's more than lactose

Kids consume a lot of carbohydrates



1. Table 1. Nutrient Intakes from Food and Beverages: Mean Amounts Consumed per Individual, by Gender and Age. *What We Eat in America*, NHANES 2015-2016. U.S. Department of Agriculture, Agriculture Research Service Website. www.ars.usda.gov/ARSUserFiles/80400530/pdf/1516/Table_1_NIN_GEN_15.pdf. Posted 2018. Accessed December 17, 2019.
2. Average Daily Intake of Food by Food Source and Demographic Characteristics, 2007-2010. U.S. Department of Agriculture, Economic Research Service Website. www.ers.usda.gov/data-products/food-consumption-and-nutrient-intakes/. Last updated June 27, 2014. Accessed December 17, 2019.
3. Dairy Products: per Capita Consumption, United States (in pounds per person). U.S. Department of Agriculture, Economic Research Service Website. www.ers.usda.gov/data-products/dairy-data/. Last updated September 4, 2019. Accessed December 17, 2019.
4. Nutrition Data System Research (NDSR) 2018 Nutrients per Food Report. University of Minnesota, Nutrition Coordinating Center (NCC). Accessed December 17, 2019.
5. Calculate Weight of Generic and Branded Foods per Volume. Aqua-Calc Website. www.aqua-calc.com/calculate/food-volume-to-weight. Accessed December 17, 2019.

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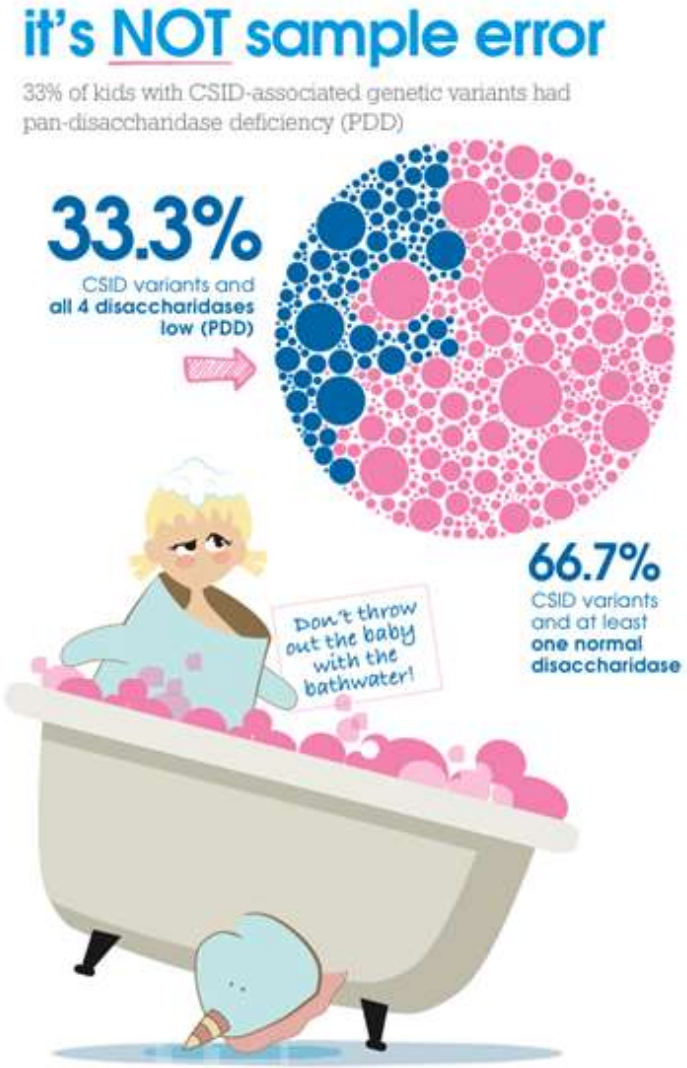
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY



Deb C, Mehta D, Ruiz V, et al. High Carrier Frequency of the Four Most Common Congenital Sucrase-Isomaltase Deficiency Pathogenic Variants Detected in Pediatric Cases with Symptoms and Low Sucrase Versus Controls with Normal Sucrase. Poster presented at: 2017 North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN); Nov 1-4, 2017; Las Vegas, NV.

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HOW IS CSID DIAGNOSED?

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Long-Term GI Symptoms, Combined With:

- Evidence of deficient sucrase activity
  - Determined by EGD biopsy and disaccharidase assay
    - 2 to 4 extra distal duodenal biopsy samples
    - Send to specialty disaccharidase testing lab
- Other tests that aid in diagnosing sucrase deficiency include:
  - ^{13}C-sucrose breath test
  - Sucrose hydrogen-methane breath test
  - Sucrose challenge symptoms test
  - Short therapeutic trial of Sucraid® (sacrosidase) Oral Solution

EGD = esophagogastroduodenoscopy

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

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Disaccharidase Assay – The Gold Standard

- Disaccharidase assay
  - Measures activity of 4 enzymes: lactase, sucrase, maltase, palatinase (isomaltase)
  - 2-4 extra distal duodenal biopsies (best near ampulla of Vater)
  - Freeze and dry ice ship to specialty lab (not path lab)
- Disaccharidase assay can be broadly helpful to:
  - Help rule in/out celiac disease or lactose intolerance
  - Assess small bowel health

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Disaccharidase Assay Reference Intervals

Disaccharidase	Normal Range*
Lactase	15 – 46 U/min/g protein
Sucrase	25 – 70 U/min/g protein
Maltase	100 – 224 U/min/g protein
Palatinase	5 – 26 U/min/g protein

*Subject to change following normal range studies conducted by the lab

Hackenmueller SA and Grenache DG. Reference Intervals for Intestinal Disaccharidase Activities Determined from a Non-Reference Population. *J Appl Lab Med*. 2016;1(2):172-80.

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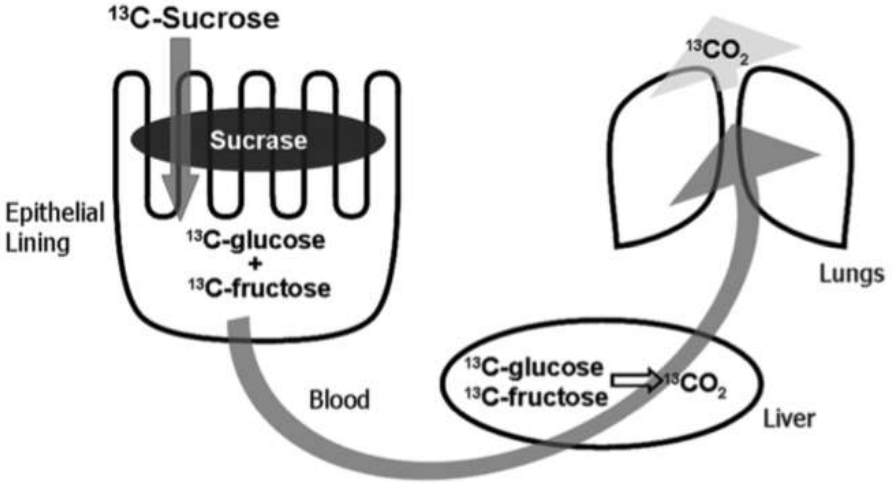
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Sucrose Breath Test

- Test is noninvasive, short in duration, and may be administered by patient in the office or at home
- Patients with CSID may experience GI symptoms from the sucrose taken for the test
- For more information, or to order a test, call 1-800-705-1962



Principle of ^{13}C Breath Test

Metabolic Solutions. Posted April 2014. www.metsol.com/wp-content/uploads/2014/04/Sucrose-Breath-Test.pdf

TREATMENT

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Dietary Changes: Low-Sucrose Diet

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Sucrose Content

Category	Food	Portion	Sucrose (grams)
Juice	Orange juice	4 oz	5.0
	Grape juice	4 oz	0.1
Fruit	Peaches, canned	½ cup	4.5
	Watermelon	1 cup	1.8
	Mandarin orange, canned	½ cup	1.7
	Banana	½ banana	1.4
	Apple sauce	½ cup	1.2
	Pears, canned	½ cup	0.7
Vegetables	Corn, canned	½ cup	3.2
	Sweet potato, mashed	½ cup	2.9
	Carrots, raw	½ cup	2.2
	Broccoli, cooked	½ cup	0.2

Created with Nutrition Data System for Research© (Regents of the University of Minnesota, 2019).

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Starch Content

Category	Food	Portion	Starch (grams)
Vegetables	White potato, mashed	½ cup	17.9
	Black beans, canned	½ cup	13.2
	Chickpeas, canned	½ cup	11.3
	Sweet potato, mashed	½ cup	9.0
Grains	Brown rice	½ cup	25.0
	White rice	½ cup	18.2
	Quinoa	½ cup	16.0
	Oatmeal	½ cup	12.7

Created with Nutrition Data System for Research© (Regents of the University of Minnesota, 2019).

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

FODMAP & CSID

Is your low-FODMAP diet still causing you discomfort? Have you considered **CSID (Congenital Sucrase-Isomaltase Deficiency)**?

Fruit
- Banana
- Blueberry
- Boysenberry
- Cantaloupe
- Cranberry
- Grapes
- Grapefruit
- Honeydew melon
- Kiwi
- Lemon
- Lime
- Mandarin
- Orange
- Passionfruit
- Pineapple
- Raspberry
- Rhubarb
- Strawberry
- Tangelo

Vegetables
- Alfalfa
- Bamboo shoots
- Bean shoots
- Beans (green)
- Bok choy
- Carrot
- Celery
- Chives
- Cucumber
- Endive
- Ginger
- Lettuce
- Olives
- Parsnip
- Parsley
- Potato
- Pumpkin
- Red capsicum (bell pepper)
- Silverbeet
- Spring onion (green section)
- Spinach
- Squash (this may be troublesome for some; check individual tolerance)
- Sweet potato
- Taro
- Tomato
- Turnip
- Yam
- Zucchini

Grain Foods

Cereals
- Gluten-free bread or cereal products

Bread
- 100% spelt bread

Rice

Oats

Polenta

Other
- Arrowroot
- Millet
- Psyllium
- Quinoa
- Sorgum
- Tapioca

Milk Products

Milk
- Lactose-free milk*
- Oat milk*
- Rice milk*
- Soy milk*

*check for additives

Cheeses
- Hard cheeses
- Brie
- Camembert

Yogurt
- Lactose-free varieties

Ice Cream Substitutes
- Gelati
- Sorbet

Butter Substitutes
- Olive oil

Other

Tofu

Sweetener

Artificial sweeteners not ending in '-ol'

Glucose

Sugar* (sucrose)

Honey Substitutes
- Golden syrup*
- Maple syrup*
- Molasses

*small quantities

This is a list of foods that are allowed on a low-FODMAP diet. The highlighted foods are high in sucrose and/or starch and may be causing your continued GI symptoms.

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

QOL Medical, LLC

Manufacturer of Sucraid® (sacrosidase) Oral Solution, an enzyme replacement therapy to relieve the symptoms of Congenital Sucrase-Isomaltase Deficiency (CSID)

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

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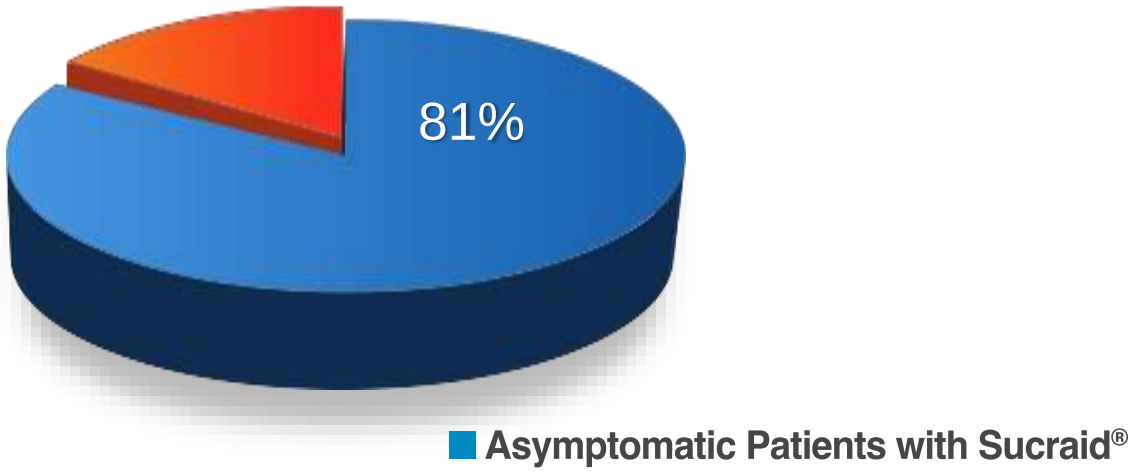
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Sucraid® Overall Symptomatic Response

81% of patients became asymptomatic* with Sucraid® in a 10-day clinical trial
N = 28



*Asymptomatic defined as symptom-free for at least 7 of the 10 study days.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

Treem WR, McAdams L, Stanford L, et al. Sacrosidase Therapy for Congenital Sucrase-Isomaltase Deficiency. *J Pediatr Gastroenterol Nutr.* 1999;28(2):137-42.

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Indication

- **Sucraid® (sacrosidase) Oral Solution** is an enzyme replacement therapy for the treatment of genetically determined sucrase deficiency, which is part of Congenital Sucrase-Isomaltase Deficiency (CSID).

Important Safety Information for Sucraid® (sacrosidase) Oral Solution

- **Sucraid® may cause a serious allergic reaction**. Patients should stop taking Sucraid® and get emergency help immediately if any of the following side effects occur: difficulty breathing, wheezing, or swelling of the face. Care should be taken when administering initial doses of Sucraid® to observe any signs of acute hypersensitivity reaction.
- Do not use Sucraid® with patients known to be hypersensitive to yeast, yeast products, papain, or glycerin (glycerol).
- Although Sucraid® provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase.

Please see additional Important Safety Information in this presentation.

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Important Safety Information for Sucraid® (sacrosidase) Oral Solution (continued)

- Adverse reactions as a result of taking Sucraid® may include worse abdominal pain, vomiting, nausea, diarrhea, constipation, difficulty sleeping, headache, nervousness, and dehydration.
- Before prescribing Sucraid® to diabetic patients, the physician should consider that Sucraid® will enable sucrose hydrolysis and the absorption of those hydrolysis products, glucose and fructose.
- The effects of Sucraid® have not been evaluated in patients with secondary (acquired) disaccharidase deficiency.
- DO NOT HEAT SOLUTIONS CONTAINING SUCRAID®. Do not put Sucraid® in warm or hot fluids. Do not reconstitute or consume Sucraid® with fruit juice since the acidity of the juice may reduce the enzyme activity of Sucraid®. Half of the reconstituted Sucraid® should be taken at the beginning of the meal or snack and the other half during the meal or snack.
- Sucraid® should be refrigerated at 36°F - 46°F (2°C - 8°C) and should be protected from heat and light.

Full Prescribing Information was provided prior to this presentation, can be accessed online at sucraid.com/pi.pdf, and is available at this presentation.

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HOW TO TAKE SUCRAID® (sacrosidase) ORAL SOLUTION

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Measure

Measure your dose with the measuring scoop provided. Do not use a kitchen teaspoon. Other measuring devices will not measure an accurate dose.

Mix

Mix dose in 2 to 4 ounces of water, milk, or infant formula. Sucraid® should not be dissolved in or taken with fruit juice.

Half and Half

For Sucraid® to be most effective, half of your dosage must be taken at the beginning of each meal or snack and the remainder of your dosage must be taken during the meal or snack.

Sucraid® dosing: < 33 lbs → 1 mL and > 33 lbs → 2 mL. It should not be mixed with hot beverages, fruit juice, or other acidic beverages as this may reduce the efficacy of Sucraid®.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

CLINICAL VIGNETTES

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Nicholas was born FT, 7 pounds and 12 ounces, uncomplicated labor and delivery*
- Exclusively breastfed; yellow, seedy stools
- Weight by 5 months: 17 pounds
- Solids started, cereals, followed by veggies and fruits
- Stools turned loose and green

*This is a real patient case study.

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Initial visit with pediatrician: viral gastroenteritis
- Support measures initiated, food stopped, and symptoms improved
- 6 months of age: solids restarted, diarrhea reappears
- PE: fussy infant, with mildly bloated abdomen, and burned perianal area
- Multiple stool samples collected, negative
- *Saccharomyces boulardii* probiotic started, some improvement noticed
- Symptoms keep on recurring

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Referral to ped gastro done by 7 months, barely any weight gain
- CBC, CMP, celiac panel negative
- Stool DNA probe for bacteria and parasites negative
- Mom noticed worsening of symptoms with fruit
- EGD with biopsy and disaccharidases ordered

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Endoscopy is visually normal
- Biopsies showed nonspecific duodenitis with no villous blunting
- Disaccharidase analysis showed:
  - Lactase 6 (15-45)
  - Sucrase 15 (25-70)
  - Maltase normal
  - Palatinase normal

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Why are both lactase and sucrase low?
- Why did he have mild improvement on *Saccharomyces boulardii*?
- What is your next step in treatment?
- Besides sucrose, which other foods can trigger symptoms?

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 1: Nicholas, Michelin Man No More…

- Nicholas was diagnosed with CSID, treatment started ASAP
  - Sucraid® (sacrosidase) Oral Solution – 8,500 units = 1 mL = 28 drops
  - Mixed in 2 oz of water or room temperature formula with each meal or snack
  - Why not warm? Why not juice?
- Reduced sucrose/maltose diet
- Immediate resolution of diarrhea. Nicholas started thriving again!*

*Results may vary.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

- Christina is a beautiful 10-year-old girl previously healthy*
- Brought directly to pediatric GI due to vague abdominal pains
- Symptoms (bloating, abdominal pain, distension, and increased flatulence) on and off for "too long" according to mom
- PE shows a normal to thin child
- Profile view shows no rib cage drop

*This is a real patient case study.

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

Further questioning:

- Mom mentions she is embarrassed some days to wear her ballet bodice due to distension
- Somewhat increased flatulence noticed
- Most abdominal pain occurs after meals

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

- CBC, CMP, Sed Rate, CRP, celiac panel normal
- Stools for ova and parasite, *Giardia* antigen, negative
- KUB: non-obstructive bowel gas pattern
- Increased intestinal air

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

Sucrose breath test:

- Baseline..... 4 ppm H_2
- 30 minutes..... 7 ppm H_2
- 60 minutes.....18 ppm H_2
- 90 minutes..... 45 ppm H_2

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CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

- Would you recommend EGD with biopsy/disaccharidases?
- How reliable is breath testing?
- Hydrogen breath test vs. ^{13}C-Sucrose Breath Test?
- How would you treat her at this point?

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Case 2: Christina, the Ballerina

- Christina was diagnosed with CSID and started on Sucraid® (sacrosidase) Oral Solution
  - Meals: 17,000 units = 2 mL = 56 drops
  - Snacks: 8,500 units = 1 mL = 28 drops
- Reduced sucrose diet
- Christina's abdomen was significantly less bloated, she had no more abdominal pains, and she was very happy!*
- She tolerates starches well, with no symptoms with maltose-containing foods (pancakes, bread, cereals)
  - Why? Because there are four different maltase enzymes, so complete maltose intolerance is extremely rare.

*Results may vary.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information within this presentation and full Prescribing Information provided with this presentation. Sucraid® may cause a serious allergic reaction.

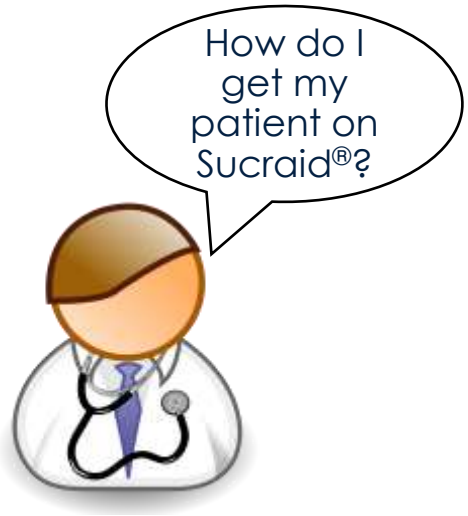
QOL Medical

PRESCRIBE SUCRAID® (sacrosidase) ORAL SOLUTION

COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

A Positive CSID Patient: What Now?

- US Bioservices is the exclusive distributing specialty pharmacy for Sucraid® (sacrosidase) Oral Solution
- To prescribe Sucraid® and minimize treatment delays, prescribing healthcare providers should submit the following:
  - Valid prescription
  - Patient's pharmacy insurance information
  - Diagnostic test results
  - Supporting clinical documentation
    - ✓ ICD-10 diagnosis code
    - ✓ Progress notes
    - ✓ Tried and failed therapies
    - ✓ Diet modifications



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COULD IT BE CSID
CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

How Do I Send in the Prescription?

Phone: 1-833-800-0122 Fax: 1-866-850-9155

HOW TO GET SUCRAID® (sacrosidase) ORAL SOLUTION

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

US Bioservices

AmerisourceBergen

Sucraid® must be kept cold and is only available from one specialty pharmacy, US Bioservices. It cannot be ordered from retail pharmacies.

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COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

Helpful Information

For Product Information:

U.S. Bioservices, Specialty Pharmacy

Phone: 1(833) 800-0122

Fax: 1(866) 850-9155

Sucraid.com

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QUESTIONS?

COULD IT BE CSID

CONGENITAL SUCRASE-ISOMALTASE DEFICIENCY

THANK YOU

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